HELPING EASE THE PAIN OF SICKLE CELL DISEASE

Sickle cell anemia is most common in western and central Africa, afflicting approximately 25% of the population. According to data compiled by the Sickle Cell Association of America, 1,000 babies are born with sickle cell disease each year in the United States. In Nigeria, 45 to 90,000 babies with sickle cell disease are born each year.

According to the United States Food and Drug Administration web-site, because of early detection of sickle cell anemia through neonatal screening and early treatment with penicillin and pain relievers, most humans with sickle cell anemia live into their 40 to 50s. In 30 years, treatment has improved. In the 1970s the majority of children with sickle cell anemia did not live past 18. The greater the number of pain crises per year, the greater their chance of dying prematurely because pain crises and oxygen deprivation damage the body's organs. Humans with 3 or more pain episodes a year usually only live to about the age of 35.

Sickle cell anemia is a homozygous genetic disease in which babies inherit a copy of the sickle cell gene from each parent. This gene is particularly common in humans with African, Spanish, Mediterranean, and Middle Eastern ancestry. In the United States sickle cell anemia is most common in humans with African ancestry.

The physiology of sickle cell anemia is posted on several web-sites; I particularly like the Mayo Clinic Website. Briefly, sickle cell anemia is a disease of red blood cells. Red blood cells are essentially packets of hemoglobin, of which the main protein is the oxygen-carrier. In healthy humans, the main form of hemoglobin is hemoglobin A. In humans with sickle cell anemia, the hemoglobin has a single change in the protein chain, and this form of hemoglobin is known as hemoglobin S. The mutation in the hemoglobin gene changes the conformation of the hemoglobin. Instead of the red blood cells being their normal concave shape, they become sickle shaped, hence the name of the condition, sickle cell anemia.

Normal red blood cells are round and flexible, allowing them to travel through narrow blood vessels. Due to the rigid structure of sickle red blood cells, they tend to stick together and block narrow blood vessels. Such blockages prevent oxygenated blood from reaching all parts of the body. Oxygen deprivation causes tissue and organ damage, stroke, and painful episodes (crises) in a patient's joints and bones. Sickle red blood cells are fragile; the body's rapid destruction of sickle cells leads to anemia, jaundice, and gallstones.

Treatments for the complications associated with sickle cell anemia include antibiotics (penicillin), pain relievers, intravenous fluids, blood transfusions, vaccination against pneumococcus bacteria, and folic acid supplementation.

The signs and symptoms of sickle cell anemia vary. While some patients have mild symptoms, others have severe symptoms that require hospitalization. Pain crises also differ in intensity, frequency, and duration. Most crises can be managed at home with pain medicines, rest, and extra fluids. However, during a severe pain crisis, patients may be hospitalized for treatment with intravenous hydration and pain medication. While 1 patient may only have a single crisis a year, others have crises more frequently. Crises may last days or even weeks.

Sickle cell anemia increases the risk for infections, stroke, and acute chest syndrome. Acute chest syndrome is caused by infection or trapped red blood cells in the lungs. When humans with sickle cell disease get acute chest syndrome, they may have severe chest and abdominal pain, fever, cough, and trouble breathing.

Sickle cells damage the spleen, an organ that helps the body fight infections. To protect against infections, sickle cell anemia is treated with antibiotics such as penicillin. Folic acid is a treatment option for children with sickle cell anemia. Folic acid is a vitamin that helps the body produce new red blood cells.

Diagnosis of sickle cell anemia at an early age is important for optimal treatment, which can result in fairly normal lives.

Care for children with sickle cell anemia calls for a specialized, rapid, and individualized approach. The Hematology Acute Care Unit (HACU) at The Children's Hospital of Philadelphia aims to provide such specialized care for chronically ill children.

Founded in 1998, the HACU is an outpatient unit used to admit and treat hematology patients whose estimated length of stay is less than 24 hours.

"Children with chronic illnesses like sickle cell anemia come back [to the hospital] a few times a year," according to Ms Wendy Seto RN, a nurse practitioner in the HACU. "The kids we see come back each time they have a fever, severe pain crisis, headache, or abdominal pain. With a unit such as this, we are able to form relationships with our kids to help make them feel more comfortable, and ultimately give them better care."

The HACU permits rapid evaluation and treatment of children with sickle cell anemia.

When a child arrives at the hospital emergency room a nurse practitioner quickly determines whether a child should be admitted to the HACU depending on the child's symptoms. After admittance, the nurse practitioner evaluates the child by obtaining a med-

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ical history, conducting a physical exam, and determining an assessment plan. The assessment plan is based on individual patient needs, and may include prescribing medications, determining laboratory tests, and ordering X-rays. Nurses then carry out the nurse practitioner's orders.

"A patient's care depends on the close communication and collaboration between the nursing staff and nurse practitioners or doctors," Ms Seto told me when I interviewed her during her duty hours.

Ms Seto described how the HACU promotes continuity of care, "Teenagers usually know what treatment options work for them, and often bring a walkman or book to occupy their attention. Younger children, however, tend to be more frightened and withdrawn.

"The 1-on-1 interaction between a patient and clinician allows relationships to form," Ms Seto says. "Being familiar with a clinician makes both the patient and parents feel more comfortable."

Pain is a real problem with children afflicted with this disease: "Some kids are more prone to pain crises, no matter what they do to prevent them."

When a child with sickle cell anemia is admitted to the HACU with pain, pain medications, including ibuprofen and acetaminophen with or without codeine and morphine in more severe cases, are administered intravenously. Once the pain is tolerated, nurses administer oral pain medications to ensure the patient continues to tolerate the pain. However, "children admitted due to pain crises are not completely pain free when they are sent home," Ms Seto notes.

Ms Seto explains that children brought to the hospital with fever are admitted to rule out acute chest syndrome and pneumococcal bacteria. And when children are brought to the hospital with headaches, "they are admitted to rule out stroke. Once these potentially fatal symptoms of sickle cell disease are ruled out, children are then discharged to treat their fevers and headaches at home."

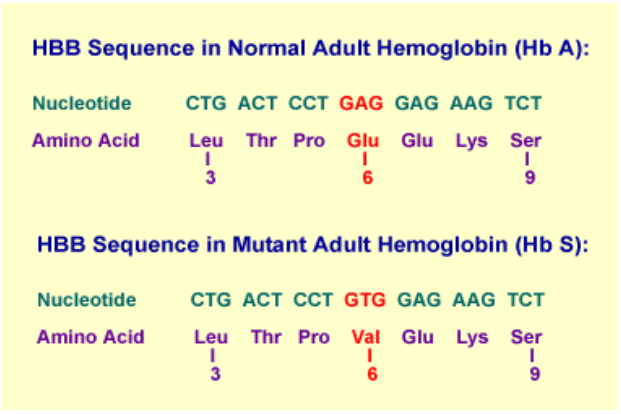
Although crises can occur unexpectedly, complications associated with sickle cell anemia can be prevented with a healthy diet, adequate hydration, moderate exercise, rest, taking vitamins and prescribed medications, and seeing a doctor regularly. At The Children's Hospital of Philadelphia, children with sickle cell disease or any known hematological condition are normally seen every 6 months in the hospital's hematology clinic.

"Parents and children receive numerous instructions and educational materials during their routine visits to the hematology clinic," Ms Seto says. "In the HACU, we try to reinforce what both patients and parents are doing right, and also what they could improve."

By EC Pierce

Website: www.sicklecellfoundation.com:

"Sickle cell disorder is by far the commonest inherited disorder in the world and 75% of cases occur in Africa. In Nigeria, where it affects 2 out of every 100 children born, it causes suffering for innumerable patients and their families. However, despite its importance, until now there has been no dedicated sickle cell center in Africa. This is partly because the very scale of the problem makes it difficult to see how to start."



Amino acid and nucleotide sequence of normal hemoglobin and sickle cell hemoglobin. From ornl.gov.

From the Nigerian Tribune 14 July 2006:

"A cure has been found for sickle cell disease and the breakthrough was made by Nigerians. The Minister of Science and Technology, Professor Turner Isoun, disclosed this at a news conference on Tuesday, July 4.....

The minister said serious research by scientists at the National Institute for Pharmaceutical Research led to the discovery. The drug, called Nicosan, was then manufactured by a pharmaceutical company in Nigeria, Xechem Pharmaceuticals Nigeria Ltd."